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Reading epilepsy: report of five new cases and further considerations on the pathophysiology

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Five new cases of reading epilepsy (RE) are reported. This is an epilepsy syndrome belonging to the group of idiopathic localization-related epilepsies. They all have some interesting features which contribute to the understanding of the pathomechanism and nosology of this specific type of reflex epilepsy. In our first patient the precipitating effect of texts in unknown languages depended upon phonematic intricacy. With our second case, changes of script within the text (Latin to Greek) increased the precipitating effect. The visual aura experiences reported in vague terms by some patients with RE may represent ictal dyslexia. For case three RE had been misdiagnosed as phobic neurosis. Even if a patient has a history of development dyslexia, and ictal dyslexia is a feature of the seizures, the onset of RE is not during primary school age but at puberty. Our fourth patient's manifestation factor of (late-onset) RE was a change of scriptural environment (Kyrillic to Latin) . Unilateral myocloni were observed with bilateral spike wave discharge in RE, and carbamazepine possibly increased the epileptic response in RE. With the co-occurrence of RE and juvenile myoclonic epilepsy in case five, the clinical features of both syndromes remained separate. All five patients responded well to treatment with valproic acid, and all confirmed that the syndrome has no tendency to deteriorate in long-term follow-up.

Key words: reading epilepsy, pathophysiology, reflex epilepsy, therapy, nosology.

INTRODUCTION

Reading epilepsy (RE) is a well-defined epilepsy syndrome which belongs to the idiopathic localization-related epilepsies. According to the International Classification of Epilepsies and Epileptic Syndromes[1], 'all or almost all seizures in this syndrome are precipitated by reading (especially aloud) and are independent of the content of the text. They are simple focal motor—involving masticatory muscles, or visual, and if the stimulus is not interrupted, generalized tonic–clonic (GTC) seizures may occur. The syndrome may be inherited. Onset is typically in late puberty, and the course is benign with little tendency to spontaneous seizures. Physical examination and imaging studies are normal but EEG shows spikes or spike waves in the dominant parieto-temporal region. Generalized spike and wave may also occur.'

For a recent review[2], 106 cases were collected from the literature. All commonly have the typical seizure pattern and precipitation is by the reading of written linguistic material. Most of them, however, have some additional features which may indicate a certain amount of variability of the clinical picture as well as contributing towards the understanding of ictogenesis.

Following two earlier case reports[3,4], five additional observations are presented here. All have some interesting details which may contribute to the understanding of the syndrome.

REPORTS OF PATIENTS

Patient 1

OH was first seen at age 15 years when he was still attending school. His family history was unrevealing; he has two older healthy siblings. At the age of 12 years he had been hospitalized for meningitis and treated with penicillin; there were no symptoms indicating encephalitis. Six months prior to admission, the day after an excessive intake of alcohol, he was reading a magazine and, for the first time, experienced frequently repeated jerks of his jaw. He was puzzled but went on reading. After about 45 minutes, his jaw suddenly became stiff, with his mouth half open. He tried to call for help but lost consciousness and fell backwards. There was nobody with him, and he awoke considerably later, feeling worn out.

Since then, the jaw jerks frequently recurred, exclu-

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sively with reading and regardless of the content. They never appeared spontaneously or when he wrote, talked or made calculations. It would start when he had read a few lines only, or later; he would then stop reading. Four months after the first seizure, again after excessive alcohol intake, he started reading and immediately had three consecutive jaw jerks and lost consciousness. His mother was with him and described a typical GTC seizure with tongue biting, respiratory arrest and cyanosis. Following this seizure, phenytoin 300 mg had been prescribed; he had a serum level of 5.9 mg/ml.

Physical examination of the patient, who was right-handed, revealed nothing abnormal. Standard electroencephaolography (EEG) with 5 minutes hyperventilation and photic stimulation was essentially normal. He was only moderately cooperative but agreed to read aloud a difficult philosophical text which led to many misreadings, repetitions and hesitations. After 10 minutes, during a period of fluent reading, he had a lightning-like perioral myoclonus which interrupted his reading for an instant; in the EEG, an irregular spike wave (SW) complex was seen on the left hemisphere with parietotemporal maximum.

One minute later, a left parietotemporal spike was accompanied by a rapid slight jerk of the jaw to the right which interrupted his reading and he refused to continue reading this text. He chose another one, in Indonesian, a phonematically easy language of which he has no knowledge. Not understanding the text, he read for 10 minutes without concentration and a lot of giggling, and nothing happened. He did not correct any misreadings during this task. During a further session, and after adminstration of 600 mg of valproic acid (VPA), he reported that he was now able to read without jerks. He read a medical text for 25 minutes with difficulty but had no jerks and no EEG discharges.

This patient was followed for 5 years at irregular intervals. He was negligent about his drugs, although he acknowledged that VPA had a very good effect. He had frequent jerks during reading, in some instances accompanied by what he reported as blurred vision. On rare occasions, he had jerks during talking, especially in vivid conversation. Twice during eating he had a prolonged feeling that his jaw was paralysed and ready to drop which reminded him of the reading seizures. He had always succeeded in preventing a tonic–clonic seizure by stopping reading when he felt the jerks.

Patient 2

PA is a 23-year-old male student of history who is right-handed. A male cousin of his mother suffered from epilepsy, and is reported to have died from a seizure whilst taking a bath. No details about this relative's epilepsy were available. At birth the patient was cyanotic after delivery but made a normal development. As an infant, he suffered from bronchial asthma which subsided at age 6–7 years.

He had his first GTC seizure at age 17 years in school; no aura was reported. He received diphenylhydantoin (DPH) from the general practitioner who transferred him to Prof Penin of Bonn for more detailed investigations. Physical and neurological findings were normal but the cranial computer tomography (CCT) was reported as demonstrating a slight cortical atrophy of the right rather than the left occipital region, and a slightly dilated right lateral ventricle. These findings were considered consistent with the history of a perinatal insult with blue asphyxia. The EEG at a DPH level of 11 mg/ml was unrevealing. However, a second investigation, a 48-hour ambulatory cassette EEG without medication, showed a total of 16 short bursts of bilateral irregular and asynchronous spikes and waves of variable laterality (Fig. 1). As at that time no indication of RE existed, it was not noted if the bursts occurred with reading. The medication was changed to VPA which was stopped after 2 years without seizures.

At age 23 years, the patient had another GTC seizure which prompted renewed VPA medication. He was referred to our clinic and reported that this seizure had happened during reading. He had felt stressed on the day of the seizure, and had been reading a text which was of no particular significance to him but dealt with Greece and contained interspersed words written in Greek script which the patient was able to decipher with some effort. After reading for several minutes, he found it difficult to go on because, repeatedly, even if he still could see the letters, he could no longer recognize the words which they composed. He tried to read on but had repetitive moments of alexia, and, after 1–1.5 minutes, lost consciousness. He further reported that he had previously experienced similiar paroxysmal alexias during reading of texts with intricate terminologies. This occurred on three or four occasions during each of which he had interrupted his reading. He remembered that one text was early medieval, in Althochdeutsch which is quite different from contemporary German.

Reappraisal of his first seizure revealed that this had occurred in his French class when, having been inattentive, the teacher had made him read a French text aloud. He was taken by surprise and was unsure where to begin. In this situation of emotional stress the seizure occurred after the first few words.

Neurological findings of the highly intelligent, cooperative patient were again normal. Routine EEGs including hyperventilation, intermittent light stimulation and some sleep stage I, taken with a VPA dose of 600 mg (serum levels around 50 μg/ml) showed some slight non-specific abnormalities such as temporal dysrhythmias with left preponderance, bilateral parieto-occipital small steep waves, and a 1 second group of

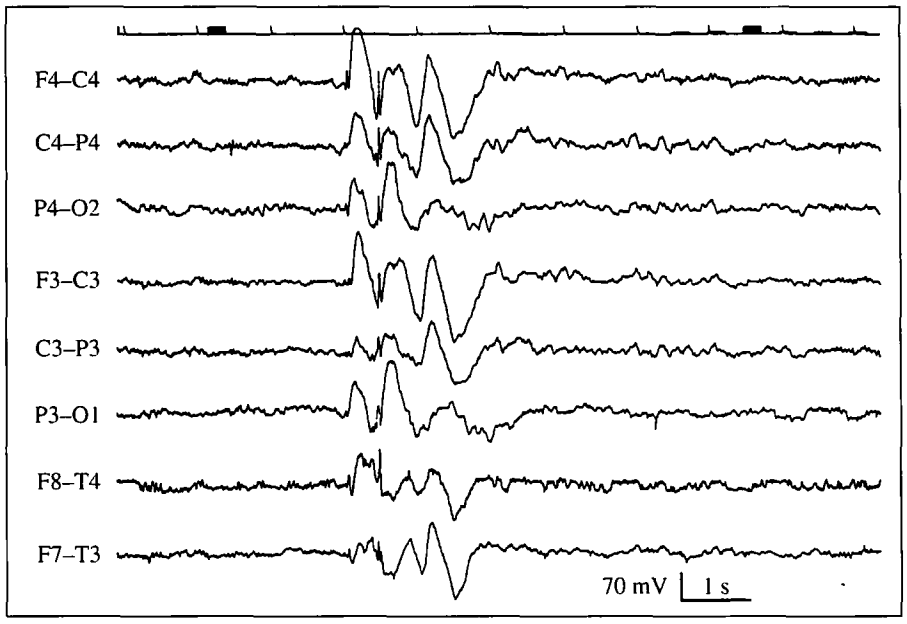

Fig. 1: Spontaneous spike and wave discharges in the 24-hour EEG of pt 2. The slightly asymmetric discharges of changing laterality are here accentuated on the right. (Courtesy of Professor emeritus Dr Heinz Penin, Bonn.)

rhythmic 2.5 Hz 50 mV waves after 5 minutes of hyperventilation.

Regarding the rarity of the events and the patient's hesitation about suppression of medication for diagnostics, no reading EEGs were taken. The patient had to read intricate material in his professional life and felt that he could not rely on getting a warning early enough to avoid a GTC seizure, so he preferred to pursue pharmacotherapy. The VPA dose was slowly withdrawn over 4 years, starting 2 years after the last seizure, and he remained symptom-free. He works now as a newspaper journalist and takes care not to read for extended periods, especially when linguistically intricate texts are involved.

Patient 3

HB is a 35-year-old male mechanic, he is married and has a healthy 7-year-old son and a negative family history. The patient is right-handed and has had no major physical illnesses, but reports his education and childhood development as problematic, being characterized by demanding and punishing parents. It seems that he had some difficulty with learning to read, and his mother often had him read aloud and would hit him at misreading. Soon, reading became to him a source of deep anxiety. Until young adult age, he suffered from nocturnal enuresis.

When he was 13 years old, he had his first GTC-seizure with deep cyanosis. This occurred in school when he was asked to stand up and read aloud. He felt very uncomfortable to do this standing in front of his class, but began reading with increasing stuttering. He could not pronounce the last word but had a funny feeling in his throat like a hiccup, and his tongue seemed to become thicker. He felt a tension in his head, and his face felt blown up. The last feeling before losing consciousness was like suffocating.

Since then, he would always have similar feelings in his throat and tongue when reading, and he had consequently restricted his reading to the minimum possible, because he was afraid to have another seizure. When an aura developed, he would stop and look away. He was able to keep control but the auras would reappear after a short time. At 18, 20 and 31 years, three additional seizures with loss of consciousness and slow recovery occurred, all with silent reading. Typically the more intense auras were accompanied by an impossibility to grasp the words before him as words, although he still saw them as visual patterns.

He especially avoided reading aloud, and he had observed that he must avoid eating during silent reading because this would precipitate auras just as much as reading aloud. On rare occasions (about five times a year) he would also have an aura when talking rapidly especially if he felt nervous or emotionally affected in special situations.

For the last 2 years, his anxiety had been increasing to panic-like episodes accompanied by flushes, not only when he had to read but also when confronted with other people. For this he had been given bromazepam which stopped the auras during reading but did not improve the anxiety so that he stopped intake after a week, and the auras reappeared. Shortly before we met him, he had been hospitalized with a diagnosis of phobic neuro-

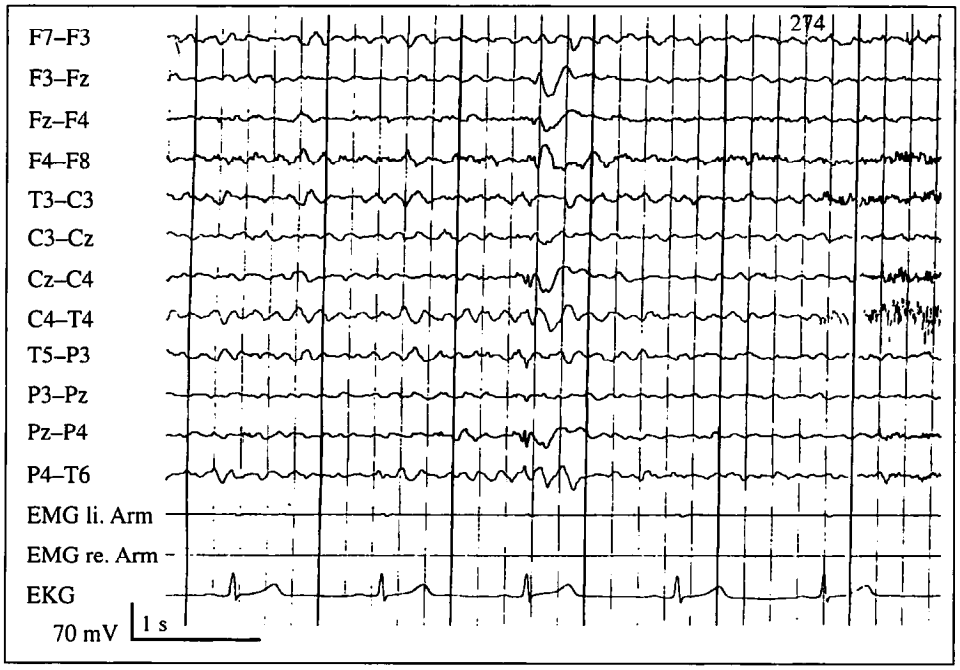

Fig. 2: Patient 3 reading aloud. Towards a background of rhythmic temporal theta activity of right accentuation, a perioral myoclonic jerk is accompanied by a miniature sharp wave with exceptionally left centroparietal parasagittal phase reversal.

sis or reading phobia. He had received psychotherapy, and benefited from it regarding his anxiety, but he had also become more calm about reading and had occasionally been able to read for a while. However, the feelings in his throat which made him stop reading had not disappeared.

We found a physically healthy but psychically depressed and distrustful patient whose routine EEG, including 5 minutes of hyperventilation and intermittent light stimulation, was unrevealing. He was at that time unmedicated. His CCT showed slight signs of left temporal atrophy.

After some hesitation, he consented to a reading test with video-EEG recording. This lasted 2 hours including a meal break of 30 minutes and comprised silent and loud reading of a political newspaper article, a technical text from his professional field, a Swiss detective story (i.e. a German text with unfamiliar idioms), and an English text (the patient reads English with difficulty and many interruptions).

Even with the professional material, which he understood easily, his performance was imperfect with many interruptions, even at common words and simple figures. He started to read aloud, and, after 2 minutes, began to report aura feelings in his throat, part of which were accompanied by a slight rapid pouting of the lips, which would interrupt him in the middle of a a word. With silent reading no visible signs occurred but he indicated the auras which were less frequent. In the EEG ictal sharp waves and steep transients were observed mainly in the right central and mid-temporal regions.

Prolonged reading caused anxiety and sweating. Both the English and the Swiss texts were difficult for him to pronounce, and this brought him repeatedly to a complete stop. A typical example was the following sentence: 'Er war barhaupt, sein Haar zurückgeschnitten, vom Hinterkopf stand eine Strähne ab, wie die Feder bei einem Reiher', which is a somewhat unusual description of a person's appearance. He read: 'Er war...(pause)...er war...überhaupt kein ...(pause) sein Haar...(long pause) Er war behaupt, sein Haar...(long pause, grimaced, drew the book nearer to himself) zurückgeschnitten, vom Hinterkopf stand eine (myoclonic jerk)... eine Strähne ab... wie (long pause) die Feder von einem Reiher.'

The patient reported afterwards that he had not understood the meaning. He had one myoclonic seizure during the test at the end of this sentence, and another one immediately in the following whereafter he declined to go on reading.

The patient was given CBZ which, at a maximum level of 10.6 μg/ml, had no influence on the condition. The medication was then changed to VPA monotherapy of 1200 mg, and his auras disappeared completely. He has now been on this medication for 6 years with excellent seizure control. Only when he forgets to take his medication for two subsequent days will he have his habitual speech motor seizures. He now reads much more, without anxiety, and seems to enjoy it although he is still less interested in books than in journals and letters. He has received psychotherapy for nearly 1 year, which has relieved his panic attacks. He has had a full-time

job for several years.

A recent control EEG, with reading aloud for during nearly 2 hours, showed right temporal and central steep transients. His reading of texts about psychoanalysis and about computer software was not very fluent. He had three short motor auras and interrupted reading; however, due to his increased confidence in his medication, he was not afraid of having a seizure with loss of consciousness and he continued the test. With 1200 mg VPA per day his serum level was 67.4 shortly after the EEG recording.

Patient 4

AG, a 24-year-old married turner, grew up in a germanophonic community in Kasakhstan where he lived until the age of 21. The family language was German but the school language was Russian, and all books were in Kyrillic script. He only learned the Latin alphabet after he had moved with his family to Germany. He is intelligent and hard-working, and has adapted very well to his new surroundings. Toward the end of his first year in Germany, he experienced jaw jerks for the first time when silently reading a German newspaper. These became increasingly frequent and occurred with both loud and silent reading of German and Russian texts. When he read rapidly, jerks were more likely to occur.

After approximately 1 year, in a period of repeated lack of sleep, he had his first GTC seizure after 10 minutes of reading a newspaper, starting with a tonic contraction of the jaw and a cry which he did not remember. One year later, again following sleep deprivation, he suffered another two GTC seizures at a short interval, both precipitated by reading and preceded by oral twitchings. This prompted admission to our hospital with a suspicion of epilepsy.

He is the fourth of a family of eight, and there is no family history of epilepsy. His own history is unremarkable, apart from a fracture of the jaw at age 20 which he at first believed to be responsible for the jerks. He is right handed, and his physical examinations were normal. He was unmedicated, and the routine EEG showed some steep theta waves during hyperventilation; he is not photosensitive.

He underwent a series of reading tests with EEG and EMG of mental muscle or masseter. The German reading matter comprised various easy texts, a nonsense text, a fairy tale, a drama by Dürrenmatt where he tried to differentiate the parts of the dialogue, and an unfamiliar technical text about EEG machines. In Russian (Kyrillic script) he read a journal article of no particular interest, another article about political prisoners, and poems by Pushkin. He also read rows of figures as well as the same figures spelt out in letters, scanned a series of traffic signs and explained their meanings, and sang songs in German and Russian.

The texts were presented in different sizes, both binocularly and monocularly left and right. He read both silently and aloud, slowly and rapidly. All tests were video documented. He was fully cooperative, and demonstrated fluent reading skills with both types of script.

Part of the tests were done unmedicated, partly with CBZ (with levels rising to 12.4 mg/ml), which had no clinical effect, and partly after addition of VPA 600 mg which considerably improved the condition. A final control test with VPA 1200 mg/day as monotherapy (trough level 81.6 mg/ml) was essentially unprovocative (see below).

All linguistic material proved provocative, and there were no clear differences between both languages, contents or size of text, binocular or monocular presentation. Loud reading was clearly more provocative than silent reading, and fast reading more than leisurely reading. He noticed that he felt uncomfortable with the unfamiliar investigation procedure. The typical provoked event was a single, rapid jerk of the jaw to the right. In early test phases, the jerks were preceded by subjective feelings of some change in the jaw, and on rare occasions, a slight rapid contraction occurred in the right angle of his mouth. All events were accompanied by steep theta transients, sharp waves or sharp and slow waves which were restricted to or most prominent in the left centro-anterior-temporal region (Fig. 3). Not all EEG events were accompanied by clinical events, and with prolonged reading they increased in frequency. On one occasion (unmedicated), after prolonged reading of a German newspaper, shifting between loud and silent reading, the EEG events reached a density of about one every 5 seconds, and the patient asked to finish the test because he feared he was going to have a seizure if he continued.

During a 10-minute reading of pictograms (traffic signs) a subclinical steep transient in an EEG was observed where linguistic material was very provocative. A reading period was sometimes followed by some steep transients when at rest. Reading of figures in both presentations, reading of a dramatic dialogue and singing were unprovocative in a test session after administration of VPA with findings greatly decreased. However, EEG events could still be evoked by prolonged continuous reading of German and Russian, both silently and aloud, especially with rapid reading.

Misreading occurred in texts that were difficult to read, and such material was more provocative. The misreadings, however, did not immediately provoke jerks or sharp waves. The jerks, on the other hand, seemed not to interfere with the reading any more than by the momentary muscular action; they did not impair his perception of the texts.

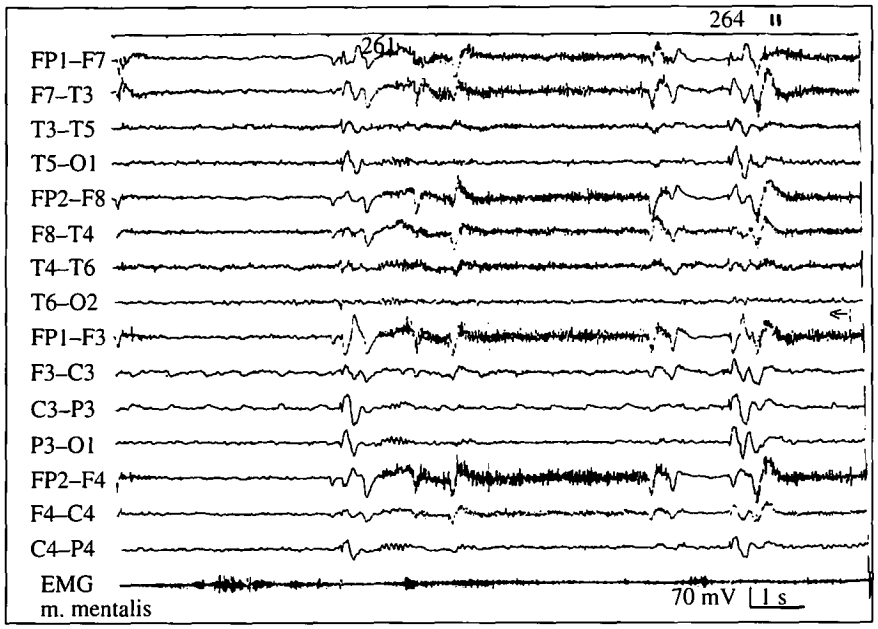

Fig. 3: Patient 4 reading aloud (see activation of mental muscle). Myoclonic jerks of the right angle of his mouth are accompanied by relatively widespread single 3 cm/second spike waves on the left hemisphere, with fronto-temporo-central maximum.

There was no positive influence on the condition due to CBZ monotherapy; the patient rather felt that he became more sensitive to reading, and also now had occasional jerks in vivid talks. On one occasion he reported a jerk when he was attentively listening to a vivid conversation. Chewing remained unprovocative. With VPA monotherapy the patient had complete clinical control, and was now able to read for long periods without any jerks or strange feelings in the jaw. In the EEG, only rare left-temporal steep transients were now seen, but no sharp waves.

Patient 5

JS is a 47-year-old female laboratory technician without familial antecedents who has had two GTC seizures in her life time. The first was at age 17 years during an operation on a nasal septum which had to be interrupted. The second occurred at the age of 29 shortly after another surgical procedure. Since the first GTC seizure, she occasionally had bilaterally myoclonias of the arms 'as if a birds beats its wings', in full consciousness. These occurred more readily after increased alcohol intake or with prolonged TV watching. These myoclonias were treated intermittently with benzodiazepines which she took if feelings of unrest or tension of the neck indicated to her an increased probability of having jerks. After her second GTC seizure an EEG was taken and DPH was prescribed which she took for several years.

In that period she first noticed that, often with continuous reading, she would develop jaw jerking. This would occur after reading about five pages of unspecified texts. She found, however, that more difficult texts, reading English or reading aloud were more provocative. With reading aloud, she was unable to go on, not only because of the jerks but she could no longer form words from what she saw. It was as if she could no longer grasp the reading material, although she still saw it clearly.

These symptoms occurred more readily when she felt exhausted, distressed or with lack of sleep. If jaw jerking occurred, she paused, and the jerking would immediately stop. If she had to read or wanted to read more, she took 10 mg oxazepam and knew that 30–45 minutes later she would be able to read without difficulty. However, she gave up her former habit of reading a whole night through if she had a book which interested her highly.

Over the years these symptoms had slowly increased and jaw jerking would now also occur in emotional discussions, especially when she was in a fury or when she felt unusual or distressed. Occasionally, when writing a long letter, she would have jerks in her right arm, sometimes together with jaw jerks. During writing she found these jerks to be clearly different from the above-mentioned bilateral myoclonias. The latter had never evolved out of the jerks which were provoked by reading, talking, and writing.

Her past history was otherwise normal, and her physical examination unrevealing. A CT scan at age 32 had been described as showing slight cortical atrophy.

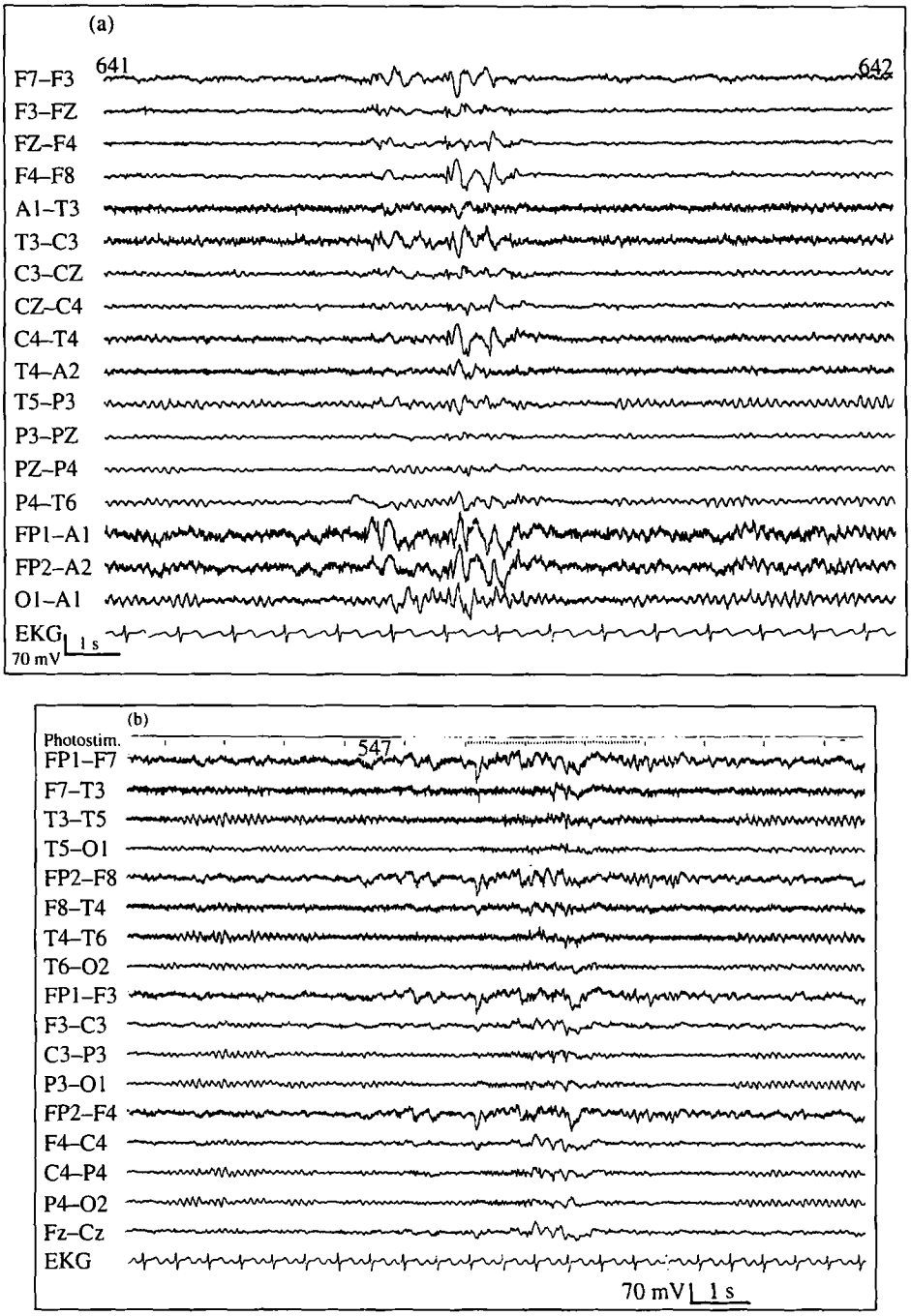

Fig. 4: (a) Patient 5 at hyperventilation. Short subclinical volley of irregular bilateral 3 cm/second spike waves, here shifting from left to right. (b) In the same EEG, at 20 Hz photic stimulation, about 800 milliseconds after eye closure, following a phase of photic driving, a short group of miniature bilateral spike waves is registered (medication: phenytoin 300 mg).

A routine EEG, including 3 minutes hyperventilation and photic stimulation, was taken with a medication of 300 mg DPH. This showed rare subclinical irregular rapid spike waves at rest, with hyperventilation, and with photic stimulation, where they could be precipitated by eye closure (Fig. 4).

Unfortunately, the patient did not consent to a reading test at her first investigation. Valproic acid was prescribed which had the remarkable effect that, from the first tablet, she clearly felt this was a drug which could help her, and she has indeed never since had any provoked or unprovoked myoclonias. She reads now quite at her ease. As a consequence, she has become much more cooperative, and most of the details of her seizure history were volunteered after the onset of therapy. However, she still politely declines suggestions to have a reading EEG done without drugs. Thus, her ictal EEG is not known.

With a medication of 900 mg VPA she is practically seizure-free, with exceptional brachial myoclonias following lack of sleep or with antibiotic treatment for a respiratory infection.

DISCUSSION

The clinical syndrome of RE is today well established. Some details still need clarification, however, and there are some controversial points. Therefore, new cases are worth studying as they may provide useful additional information.

Thus, one controversy is about the provocative effect of foreign languages. Provocation of seizures and EEG discharges by meaningless material as in patient 3 is an important argument supporting the view that the precipitating mechanism is related to the transcoding procedures in reading rather than to semantics[4]. There are, however, patients where reading of a foreign language has been innocuous[5]. Forster criticized that these reports fail to describe the patient's performance, and points out that only those tests which the patient takes seriously can be expected to give any results. A parallel to his observation is our patient 1 who ridiculed his reading of Indonesian, made no attempt to read it correctly, but also had no difficulties with this phonetically easy language. The test was not provocative.

The importance of the formal procedure is underscored by our patients 2 and 4 which demonstrate the possible influence of the intricacies of a script in two different ways. Patient 2 had his second GTC seizure when he read a text where Latin script was interspersed with some Greek script. The bilingual patient 4 developed RE when he had to adapt from a Kyrillic to a Latin scriptural environment.

Patient 2 has only had two GTC seizures, and few isolated visual auras, but the precipitating situations in every instance are so unequivocal that we feel confident about the diagnosis. This patient, who is a keen observer, has made us understand the probable significance of the aura sensation which many patients vaguely describe as 'blurred vision' or the like. At least in this patient, and probably also in some others, such as our patients 3 and 5, it is an ictal alexia. This is by no means surprising as a close relation between provoking stimulus and seizure symptoms is also observed in other 'reflex epilepsies' (eye closure sensitivity[6]; seizures induced by movements). Likewise, the typical motor phenomena involving the muscles subserving speech have been explained by the active involvement of this musculature in the process of (even silent) reading[7]. The difference between loud and silent reading can be explained by an increase of motor activity in the speech musculature. The same effect was obtained in our patient 3, when he increased this activity by eating during silent reading. In one patient who read Braille, the jerks involved the hand used for reading[8], and the same occurs in those patients in whom writing is also provocative such as our patient 5. All these, however, are accidental symptoms, and even with the attention focused on possible alexia, it was clear in our patient 4 that he did not have it.

Patient 3 presents a history of an unusually tenacious misdiagnosis of a phobic neurosis in spite of a typical clinical syndrome. When he was first relieved of his seizures, it was interesting to see how little he had become neurotic, in spite of many good reasons to do so.

Just as patient 1, this is one of the patients with RE who are poor readers. He may even have had developmental dyslexia the role of which in the development of RE has been hypothesized previously[9]. Nevertheless, his seizures did not start at early school age but at the age typical for RE. This is important as it further demonstrates that RE has not primarily to do with reading.

In all five patients it was true that intricate texts are especially provocative. We have argued previously[2,3] that the misreadings which occur more readily in difficult texts directly precipitate seizure discharge by an overload of the systems involved in the script analysis. Although this is certainly true for some patients, it could clearly not be observed in our patients 1 and 4 when they read aloud with video control. Nonetheless, it was obvious that misreadings had some—less direct—influence in their ictogenesis. This influence may be more complex then it appears at first sight. Recent research on reading[9] has revealed that different strategies are probably used for short and common words and familiar short combinations of words than for other texts. Material of the first type seems to be processed in a holistic ('kanji') way, perhaps involving the right hemisphere much more than the latter type which is analysed more literally. Reading of difficult texts which may even result in misreadings may mean a change of strategies with involvement of different neuronal subsystems.

In this respect, it is interesting to note that the seizure discharge in RE is mostly lateralized to the left, but in a substantial minority of patients it is to the right[2]. One such example is our patient 3 who clearly followed a holistic strategy of script analysis (tried to grasp whole words) in the example given above.

In our patient 2, the laterality of the spike wave was shifting, in three others it was to the left, and in two of these it was observed that their jaw jerked to the right.

Non-specific factors increasing the risk of seizures were noted in our patients 1 (alcohol excesses) and 4 (sleep deprivation). Emotional stress seems to be a fairly common non-specific modulator in this syndrome, and could trigger seizures induced by reading,

and sometimes talking, in all patients of this series.

Several of our patients also had occasional seizure symptoms with talking, especially if there was emotional involvement (patients 1, 4 and 5), with eating (patient 1), and once with attentive listening to a vivid conversation (patient 4), an activity which may involve some amount of silent wording in response. Interestingly, this feature developed in consequence of CBZ medication which seemed to increase rather than alleviate the epileptic responses.

Patient 5 is unique because her history combines features of juvenile myoclonic epilepsy and RE (this has been observed also by Radhakrishan *et al*[10]). She was also photosensitive, and it will be remembered that photosensitivity is most frequently observed in juvenile myoclonic epilepsy[6]. From a nosological viewpoint it is important to note that the features of both syndromes remained separated although both responded equally well to VPA therapy.

They became manifest at different ages, and reading never provoked the brachial myocloni of juvenile myoclonic epilepsy whereas the jaw jerking appeared exclusively in response to reading. The same was reported by Radhakrishan *et al.*[10] who, therefore, call this combination 'co-occurrence'.

In monotherapy, VPA was the most effective treatment in all five patients. One had also responded well to a benzodiazepine (bromazepam, patient 4), whereas CBZ was ineffective in patients 3 and 4, DPH in patient 1, and a combination of DPH and a benzodiazepine in patient 5. This is consistent with the recent literature from which it clearly appears that VPA is the drug of first choice[11].

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